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Counselling the Epileptic Patient

SUMMARY

Today, most epileptics can participate freely in a wide range of activities. However, their epilepsy does create some special problems. The degree to which they participate in sports is governed by their degree of seizure control, as are their employment opportunities and driving privileges. Epilepsy does not appear to be a major stress factor in marriage as long as the spouse is knowledgeable about the condition. Epileptic women usually have normal pregnancies though their relative risks are perhaps double those for the non-epileptic population. Children of epileptic women have about four times the general population's risk of seizure but the absolute level of risk is not high. (Can Fam Physician 1983; 29:107-111).

SOMMAIRE

Aujourd'hui, la plupart des épileptiques peuvent participer librement à une grande variété d'activités. Toutefois, leur épilepsie crée en effet certains problèmes particuliers. Leur degré de participation aux sports est déterminé par leur degré de contrôle des convulsions, tout comme le sont leurs chances d'embauche et leurs privilèges de conduite d'une automobile. L'épilepsie ne semble pas être un facteur majeur de stress dans le mariage en autant que le conjoint est bien informé de cet état. Les femmes épileptiques ont habituellement des grossesses normales bien que les risques relatifs soient peut-être le double de ceux de la population non-épileptique. Les enfants des femmes épileptiques ont environ quatre fois plus de chances de souffrir de convulsions par rapport à la population générale mais le niveau de risque absolu n'est pas élevé.

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THE DISCOVERY of anticonvulsants and increasing understanding of types of seizure disorder has allowed epileptic patients today to enjoy family, recreational and employment opportunities that were not feasible 50 years ago. The physician is therefore involved in at least two functions. One is to control the seizures. The second is to advise patients how their condition may affect their lives. The second responsibility is more difficult to discharge than the first because the information on which the advice is based is often fragmentary and sometimes frankly speculative. Physicians can be conservative in their opinions when 'hard data' are unavailable, but the epileptic patient may be eager to try something new.

Sports

Most physicians encourage their patients to be active and promote participation in sports to help maintain good health. This attitude is equally important for epileptics, but certain factors must be taken into consideration.

Hyperventilation may induce an absence attack or petit mal seizure in children. Theoretically, it is a problem for someone engaging in strenuous physical activity, but it does not appear to be a practical problem. Some epileptic children may have a seizure in association with mild trauma, as in tripping and falling. This type of seizure occurs infrequently, so mild trauma is not a risk factor for most epileptics.

By using modern medications, some patients have been seizure-free for a year or more when they visit their physicians to ask about sports participation. Other patients may still be having seizures, but only during sleep or under certain conditions such as eating or listening to music (reflex epilepsy) or during menstruation (catamenial epilepsy). Only a few high risk sports would be contraindicated for such patients.

Sporting activities can be classified as carrying high, moderate or low risk of personal injury. The main concern is not the likelihood of the activity provoking a seizure, but rather the result of a seizure occurring during one of these activities. The following classification is an example:

High risk: flying, gliding, skydiving, parachuting, hang gliding, rock-climbing, scuba diving, motorcycling, boxing, whitewater canoeing.

Moderate risk: horseback riding, snowmobiling, hockey, swimming, sailing, weightlifting, bicycling, downhill skiing, fishing, skateboarding, flatwater boating.

Low risk: racquet sports, golf, curling, range shooting, recreational skating, ballgames, bowling, crosscountry skiing, jogging, wrestling.

The physician must assess the risk of the sport in question and the patient's degree of seizure control or the usual seizure pattern (nocturnal, catamenial or reflex) and then formulate his advice. Low risk sports can certainly be attempted with protection such as helmets or padding, even if seizure control is not complete.

Marriage

At one time epilepsy was classified as a mental disease, and in subtle ways the epileptic may still be unfairly regarded as 'a sort of mental patient'. Fortunately, this primitive point of view is slowly changing as information becomes available. Nevertheless, epileptic patients *do* have to contend with an unpredictably disruptive condition, and according to a World Health Organization report, as many as 25% of epileptics experience recurrent mental disturbances of varying severity.[1] However, the number of patients who could be considered severely incapacitated must be significantly smaller.

Most epileptics' marital problems are rarely related to the actual seizures, but are more frequently due to secondary emotional disorders or mental alterations such as epileptic personality changes, dementias, recurring moodiness or twilight states. In some instances, marital conflicts may be psychoreactive, for example when seizures or the threat of seizures are used manipulatively, or to avoid unpleasant situations.

Epilepsy in no way jeopardizes the legality of the marriage contract but in fairness, prospective spouses are entitled to complete information about their partner's condition (even if it is fully controlled), potential genetic implications, and effects on fertility.

While marriages in which one partner is epileptic might at first seem to be especially vulnerable to stress and frequent dissolution, there is no evidence that this is true. In fact, the opposite appears to be the case; such couples often exhibit a greater degree of mutual concern and support.

Driving

The physician must decide if the epileptic patient is competent to operate a vehicle safely.

The most common situation is that of a patient with known, controlled epilepsy who wishes to apply for a personal motor vehicle license. 'Control' is assumed when the patient has been seizure-free with or without medication for one year. The physician must also feel confident that the applicant is a responsible person who will continue to take his medication as directed and will avoid precipitating situations such as alcohol abuse and excessive fatigue. Such patients are allowed to drive only personal motor vehicles and are not permitted to operate passenger transport or heavy commercial vehicles.

Patients whose seizures have been controlled for several years may occasionally stop taking medication on their physicians' advice. If they have a seizure during this time, they may drive only when they have resumed taking the previously effective dose of medication.

It is more difficult to assess the adult who has had a solitary seizure. Automobiles have become so much a part of our lives that losing the privilege of driving can have very serious economic repercussions. At the very least, it involves major changes in mode of transportation, and at worst it may even mean giving up a job. The physician is allowed considerable discretion in these cases. If the patient has been examined by a neurologist and the diagnosis of epilepsy seems unlikely, the patient may continue to drive. If a second seizure occurs, the individual must stop driving until he or she has been seizure-free with medication for one year.

However, patients who drive passenger transport or heavy commercial vehicles must stop driving for one year after even one seizure. If no further seizures occur and if they are not taking anticonvulsant medication, it is considered safe for these patients to resume their previous driving privileges.

Discretion is also allowed in the case of adults whose seizures have been strictly nocturnal or immediately upon awakening for five years or more. If their waking EEG is normal, they can be allowed to drive a personal motor vehicle.

Adolescents and young adult patients tend to be less compliant with these restrictions. One can only hope that driver safety programs in the schools will make clear to young, inexperienced drivers the necessity of stringent regulations in the interest of public safety.

Employment

Due to the variability of seizure control and seizure pattern, it would be extremely unfair to lump together 'people with epilepsy' in giving advice on employment. If epileptic patients fulfill the criteria to hold a motor vehicle license, they should be able to operate most industrial equipment safely and be suitable for most jobs. The choice of career depends on the patient's intelligence and not on the presence or absence of epilepsy. Oversedation due to anticonvulsant medication is rarely a problem, but may affect a patient's ability to study at college level.

The physician may have to consider several problems in addition to the presence or absence of seizures. If there is associated mental retardation, employment opportunities would be limited to sheltered workshops or equivalent facilities. Some patients with mild mental retardation and seizures show various degrees of social immaturity. 'Half-way houses' train these patient in lifeskills. Panorama Manor in Vancouver would be an example. It is supervised by the Vancouver Neurological Association, and is designed to provide learning experience in a closely supervised setting. Young people spend six to 12 months in this environment, after which many of them are able to be independent and hold jobs.

Some unusual personality traits may occasionally be encountered in patients with certain forms of epilepsy. For example, patients with temporal lobe epilepsy can demonstrate such traits as excessive religiosity or compulsive behavior which may prejudice their chances of steady employment. Other epileptics may have emotional problems such as recurrent depression. On the other hand, temporal lobe epileptics sometimes have compulsive personality traits, which can be extremely valuable in routine, repetitive employment.

If the seizures are not completely controlled and if they occur during working hours, patients should not seek jobs which would place them or their coworkers at risk. These patients should avoid working at heights, or with high-powered machinery. Generally, the same patients would not be allowed to drive a motor vehicle.

Pregnancy

The physician may be asked for advice by couples who are planning to have children and women who have already become pregnant. The absolute risk to the pregnant epileptic woman and her developing child is low, although some risks are higher than for the general population.

Effect of pregnancy on epilepsy: Only a few large series have looked at this problem, and they have not been well enough controlled to provide unequivocal information. The consensus appears to be that about 50% of epileptic women will have more seizures while pregnant; 42% will show no change and about 8% will show some improvement.[1, 4, 5] The chance that seizure frequency will increase during pregnancy correlates to some degree with the frequency before pregnancy. Women who had more than one seizure per month usually had more during pregnancy. Only about 25% of women who had one seizure in about nine months experienced more seizures during pregnancy.[5] There seemed to be an increased risk of seizures in pregnancy if the woman had catamenial epilepsy (related to menstruation). Surprisingly, the occurrence of seizures during one pregnancy was not a good predictor for the frequency of seizures during subsequent pregnancies.

The increase in seizure frequency might be partly related to lower serum anticonvulsant levels. This seems to occur in pregnancy even if medication is taken regularly. However, mothers concerned about potential teratogenic risks may omit medication and morning nausea may also cause a change in schedule.

Status epilepticus can occur in pregnancy but it is rare. Two separate studies of North American populations documented a frequency of about 1:20,000 deliveries.[4] Status epilepticus in pregnancy once carried a very high risk, but with newer anticonvulsants this appears to be less of a problem. Intravenous phenytoin has been used successfully to manage some of these cases.[10]

Effect of epilepsy on pregnancy: Most of the time, the course of pregnancy is unaffected by idiopathic epilepsy; however, the incidence of vaginal hemorrhage, toxemia, abortion, gestation less than 37 weeks, premature rupture of membranes, necessity for induction of labor or intervention during labor, cesarian section, neonatal deaths and perinatal deaths are all about double the general incidence of 1-5%.[1, 5] Furthermore, fertility in epileptic women is reduced, while the rate of multiple births is increased.

Isolated gestational seizures: A seizure occurring for the first time in a pregnant woman should be considered as a symptom of some underlying disorder. Investigations should consider the possibility of infection, metabolic disturbance, neoplasm, arteriovenous malformation, cerebral venous thrombosis, eclampsia or even hysteria. If no cause is found, some neurologists would withhold treatment until the second seizure. In women with true gestational epilepsy (seizures only when pregnant), 70% had only one seizure but 30% had recurrent seizures during pregnancy.[5]

Lactation: The ability to lactate is unimpaired. Very rarely, the seizure activity increases only for the duration of lactation.

Anticonvulsants: If the mother had been taking phenobarbital for seizure control through her pregnancy, the phenobarbital level will be the same in the fetal serum as in the maternal serum. Phenobarbital is excreted much more slowly in the neonate and may remain high for up to six days following birth. Parents should be warned that some neonates will show withdrawal symptoms beginning about one week after birth (after the child is already home from hospital). Symptoms such as hyperexcitability, tremor, high-pitched cries and gastrointestinal problems can then occur; these may last as long as two to four months. In contrast, phenytoin (Dilantin) also reaches the same level in fetal serum but no withdrawal syndrome has been described.[5]

Both phenobarbital and phenytoin cross into maternal milk. Phenobarbital levels in milk are 10-30% of maternal serum levels and phenytoin milk levels are perhaps 20-25% of serum levels[5]—too low to cause significant neonatal lethargy. Unfortunately, little information is available on other anticonvulsants. Diazepam crosses the placenta and also appears in breast milk at about 10% of the maternal plasma level. Due to the drug's long halflife, the newborn may develop fairly high serum levels. Nursing mothers might possibly be able to take up to 10 mg per day of diazepam without depressing the child, but higher doses would probably result in neonatal toxicity.[1]

Occasionally children born to treated epileptic mothers will show a bleeding disorder very shortly after birth due to depletion or increased metabolism of the vitamin K-dependent plasma coagulation proteins. Prophylactic treatment with phytomenadione (AquaMephyton) 0.5-1 mg intramuscularly is recommended.[4]

Genetic Counselling

Risk of offspring developing a seizure disorder: Prospective parents are probably most anxious about the hereditary aspects of their disorder. Unfortunately, there are no completely reliable data available to calculate the genetic risk, for the following reasons:

1. Epilepsy is not a single disease.
2. Population frequency of all forms of epilepsy is about 0.5-2%, so the condition is already fairly common.
3. Inheritance usually does not follow simple genetic patterns.

In general, seizures occur about four times more frequently in children of epileptic mothers, regardless of the mother's seizure type, than in the general population, for whom the overall risk is 1-2%.[1, 5] The risk is much lower for children of epileptic fathers. If both mother and father have epilepsy, the risk to the offspring of developing seizures of some type may be as high as 20-25%.

In rare situations, genetic counselling is straightforward. Seizures can be one manifestation of a more generalized systemic disorder with a well-defined inheritance pattern. Examples would be the single gene disorders that show Mendelian inheritance, such as tuberous sclerosis or neurofibromatosis (autosomal dominant), aminoacidopathies, phenylketonuria or lipidoses (autosomal recessive), Menke's disease, Pelizaeus-Merzbacher disease

(X-linked). Other conditions show chromosomal inheritance such as trisomy 13 or 18, or deletion of the long arm of chromosome 21. These are fairly easy to differentiate from the large group of epileptic disorders. In idiopathic seizure disorders, multifactorial inheritance with environmental influence is most likely, and simple Mendelian inheritance does not apply.

Within the large group of idiopathic epilepsies, different diseases and genetically distinct entities can sometimes be differentiated by a combination of clinical manifestations and EEG patterns. Metrakos and Metrakos'[1] study of the centrencephalic EEG pattern has shown the existence of an autosomal dominant gene with an unusual penetrance. It has very low penetrance at birth, nearly complete penetrance from age 4-16 years, and declining penetrance to almost none after age 40. The offspring and siblings of patients with definite seizures and the 3-Hz spike and wave EEG pattern have a 50% chance of inheriting the gene for 3-Hz spike and wave, a 35% chance of expressing that trait in their own EEG, a 12% chance of having one or two seizures, and about an 8% chance of developing chronic epilepsy. The incidence of actual seizures in relatives is therefore considerably lower than the incidence of the EEG pattern. This pattern is best considered as a marker of genetic predisposition to seizure, but other factors and environmental influences are required for the expression of the actual seizure. Some estimates can be made for certain other seizure syndromes:

Generalized epilepsy

Absence seizures (staring spells, petit-mal) with 3-Hz spike and wave in the EEG: The proband's siblings and offspring have a 7-10% risk of developing some kind of seizure disorder and approximately 1% risk of developing absence seizures.[1] About 30-40% will show the EEG abnormality and may be considered carriers of the autosomal dominant spike and wave gene. The risk for offspring is slightly higher if the mother, rather than the father, has epilepsy.

Tonic/clonic seizures (grand-mal) with spike and wave in the interictal EEG: Risks to offspring and siblings are probably about the same as for absence seizures. Monozygotic twins have an 84% concordance rate for epilepsy. Dizygotic twins have about a 10% concordance rates, similar to the rate for other siblings.[5]

Lennox-Gastaut syndrome (generalized myoclonic astatic seizures of early childhood): This is a rare and often malignant type of seizure disorder. Siblings of the proband have 13-20% risk of developing a seizure disorder and a 40% incidence of EEG abnormalities.[1]

Partial epilepsy

Benign epilepsy of childhood with central-temporal (Rolandic) spikes: This is a rather benign seizure disorder characterized by focal and predominantly nocturnal seizures. There are no organic brain symptoms and the problem almost always subsides by puberty. This is probably due to an autosomal dominant gene with age-dependent penetrance. About 32% of siblings have similar EEG findings, and 15% of siblings have seizures with the same good prognosis.[1]

Seizures secondary to trauma or infection: These conditions in the mother do not imply any increased risk to offspring. However, a genetic predisposition is suggested even within this group. EEG abnormalities and actual seizures are more frequent in these families than in the general population. The younger the proband at the onset of epilepsy, the stronger the family history of convulsions. In general, the risk for seizures for siblings and offspring is about 3-5%, if the mother's epilepsy was acquired.[1]

Febrile seizures: These occur in 3-5% of all children aged three months to five years. They are not usually classified as epilepsy (which is usually defined as recurrent non-febrile seizures). In 40% of the families of children with febrile convulsions, there is a history of other seizure disorders; this perhaps relates to a genetic influence on the seizure threshold.

About 12-15% of these children will have another febrile seizure and the risk is greater if the child was less than 12 months old at the time of the first. The risk is much lower if the children were over age three at the time of their first febrile convulsion.

The risk of future epilepsy is increased if there is a family history of epilepsy, if the febrile seizure lasted longer than 15 minutes, if it was a focal seizure or if there were several seizures in a single day, or if the child does not have normal neurological development. With these risk factors, the chance of future epilepsy is 6-17%; with them, only about 3% of children without febrile convulsions become epileptic.[1]

Risk of offspring having a malformation or disturbance of postnatal development

Children of epileptic mothers, both treated and untreated, have a higher incidence of malformation than children of non-epileptic mothers. If a large range of major and minor abnormalities are examined, non-epileptic mothers have a 3-6% risk of having a child with a malformation, whereas epileptic mothers have a 5-11% risk.[1] The incidence of malformations with epileptic mothers who are treated is twice that of epileptic mothers who are untreated, which suggests some teratogenic effect of drugs. Confounding variables probably make it impossible to compare treated and non-treated. For one thing, the epilepsy may be more serious in the treated group.

Within the broad group of malformations, there appear to be several important distinctions:

Major malformations: These are comprised mainly of oral-facial clefts (cleft lip, cleft palate) and cardiac lesions (usually septal defects). They are seen in children of both epileptic mothers and epileptic fathers, with a very similar incidence for either parent. The risk appears to be from 2-4% for either type of malformation. The risk for children of epileptic parents is therefore 5-10 times higher than in the general population.[4]

These abnormalities are not drug-specific, dose-specific or serum level-specific, and have been reported in children whose mothers were taking the standard anticonvulsants, both in combination and separately. Furthermore, children of mothers who took phenobarbital throughout their pregnancy for reasons other than epilepsy do not have an increased risk of these malformations.[1]

Minor malformations: Minor craniofacial abnormalities such as short nose, flat broad nasal bridge, hypertelorism, epicanthal folds, antiverted nares and some abnormalities of the digits such as hypoplasia of distal phalanges and hypoplastic nails may occur in 9% of the offspring of epileptic women taking phenytoin, pheno-

barbital or primidone. It is difficult to decide whether these defects are embroyotoxic manifestations of the anticonvulsants, or related to the underlying epilepsy.[1]

Animal studies certainly support a teratogenic effect of these anticonvulsants when given at certain times during pregnancy or in high doses, although there seem to be no specific syndromes related to specific drugs.[1,2] However, if one child is born with minor craniofacial and digital abnormalities, the likelihood of subsequent children having them is higher if the mother takes phenytoin when pregnant. While phenytoin is especially implicated, the defects that occur with carbamazepine or phenobarbital are qualitatively the same. Therefore, the phrase 'fetal hydantoin syndrome' is probably inappropriately specific.[8] Similar craniofacial abnormalities have been described with trimethadione, though some of these children also have high-arched eyebrows.

Other evidence suggests that the maternal epilepsy itself is the important variable. In one study, none of the children of non-epileptic mothers showed these minor abnormalities, nor did the children of epileptic fathers.[1] Further, these abnormalities occurred to at least some degree before phenytoin or phenobarbital were widely used in treating epilepsy. The children of mothers treated with phenobarbital for reasons other than epilepsy did not show an increased incidence of malformation.

If there is risk of fetal malformation, carbamazepine appears to be the safest anticonvulsant to use during pregnancy.

Postnatal development and cerebral disorders without dysmorphic features: There appears to be no consistent difference in the birth weights or lengths of offspring of epileptic mothers in comparison to control groups. There is, however, a suggestion that head circumference at birth may be slightly reduced.[1] Perhaps 8-16% of children have a head circumference below the third percentile, a four-fold higher incidence than in the general population.

When these children are assessed at intervals of a few years, more than the expected number of children are at the lower levels of the somatic parameters of height, weight and head circumference. This is thought to be more than expected on the basis of strict genetic factors, but it does not appear to be related to the different forms of maternal epilepsy, nor to specific drug use. It tends to be seen more often in boys.

Psychomotor and intellectual development also appear to be affected to a slight but significant degree.[1] There is a trend towards lower IQ ratings at four and seven years of age, and scores for motor development are also lower. No specific anticonvulsant has been implicated, but unfortunately there is no good comparable study on children of untreated epileptic mothers. This trend towards a lower IQ may be due to postnatal factors such as poor parenting (due for example to being on sedating drugs), insufficient intellectual stimulation in the home, or inadequate nutrition.

Conclusion

The absolute frequency of problems is very low, despite the increase in relative risk. Epileptic parents need not be discouraged from having children for this reason alone. Because palatal closure occurs at about the 47th day of gestation, there would be no point in switching a patient from phenytoin if she presented in the second or third month of pregnancy.[7] Women who are not yet pregnant could consider switching to a possibly more harmless class of drug such as carbamazepine.

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